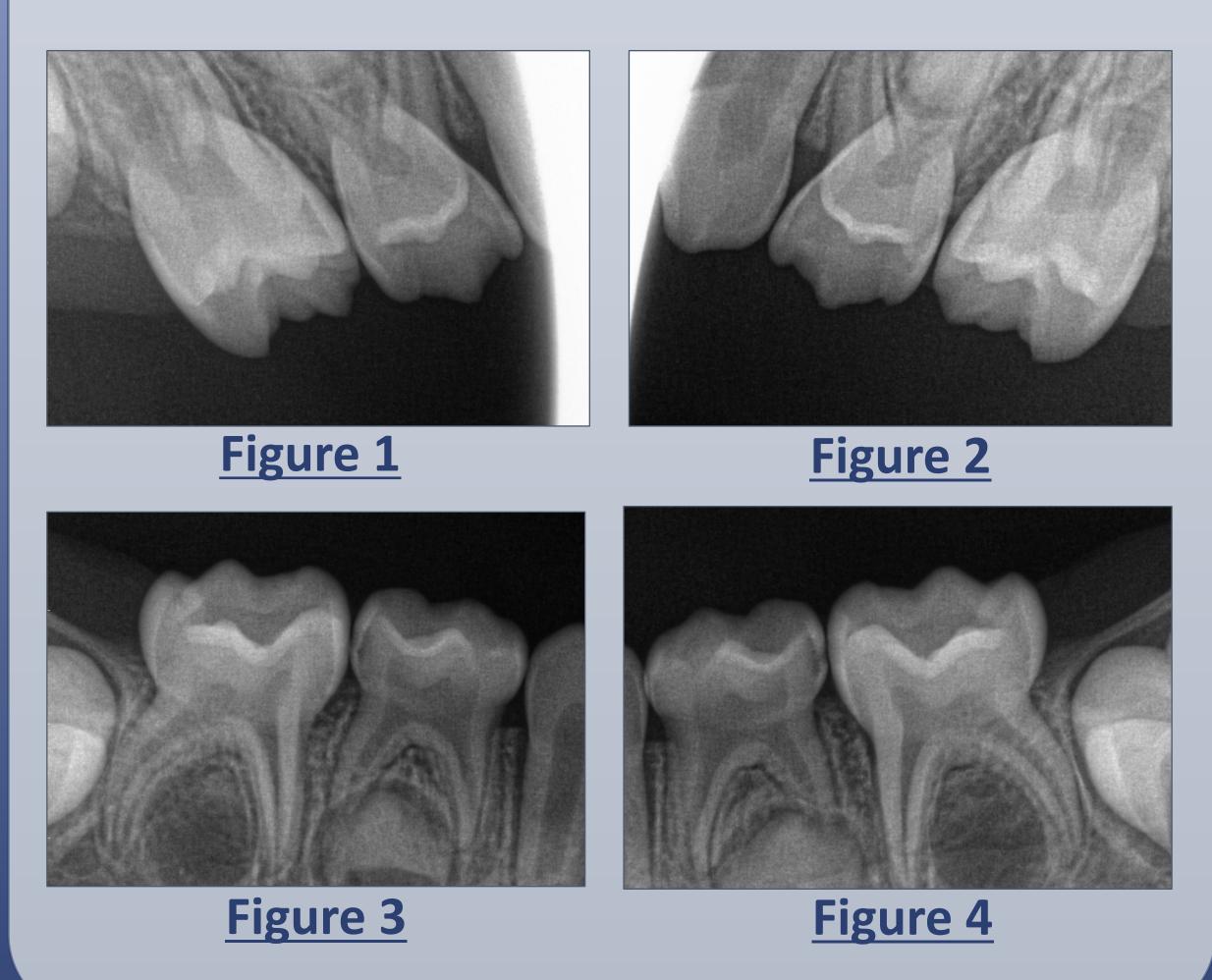


ABSTRACT/INTRODUCTION

Fanconi anemia is an inherited autosomal recessive bone marrow failure syndrome that affects proper development of white blood cells, red blood cells, and platelets¹. This eventually leads to hematopoiesis and the need for a bone marrow transplant as hematopoietic abnormalities are eventually found in almost all patients⁴. Fanconi anemia also makes patients more susceptible to malignancies, including squamous cell carcinoma of the head and neck and leukemia². This case report details a 4-year-old female patient seeking dental clearance for her upcoming bone marrow transplant due to a diagnosis of Fanconi anemia at the age of 2 after a workup for thrombocytopenia. The purpose of this report is to review dental treatment considerations for pediatric patients with Fanconi anemia prior to transplant.



Dental Rehabilitation of 4-Year-Old with Fanconi Anemia *Shelton H, Onikul R, Burleson A, Patel N, Sparks J, and Bohaty B Children's Mercy Hospital and UMKC School of Dentistry, Kansas City Missouri.

CASE REPORT

A 4-year-old female patient presented to the clinic for radiographs and an exam to obtain dental clearance for her upcoming bone marrow transplant for treatment of Fanconi anemia. The mother reported no familial history of Fanconi anemia. The patient also exhibited decreased growth, weighing only 11.50 kilograms, in the 0.15th percentile for her age, at her initial appointment. The patient had been referred to our clinic after an outside dentist recommended three stainless steel crowns to obtain dental clearance for a transplant. Initial in-office PA radiographs showed carious lesions on #B, #I, #L, #S, with no reports of pain or discomfort (Figures 1-4). Due to the patient's age and significant health history, it was planned to treat teeth with decay with stainless steel crowns. The mother opted to attempt in-office treatment for the patient due to a history of difficulties waking up from general anesthesia. Upon reporting to her hematologist, it was noted that her platelet count was 26,000. AAPD guidelines at the time required platelet counts of no less than 60,000 for invasive treatment⁵. The patient received a platelet transfusion one day prior to her dental procedure, which raised platelet levels to 113,000 1-hour post-transfusion. She was also started on an antifibrinolytic on treatment day pre-operatively to continue 2-3 days postoperatively. She presented for in-office treatment, but exhibited uncooperative behavior, so treatment was aborted. Per hematology, treatment would need to be completed within the 14-day period after platelet transfusion in order to avoid requiring a second transfusion as limiting blood transfusions in patients who need bone marrow transplants is critical. The patient was then scheduled for same day surgery 4 days post-transfusion. While under general anesthesia, more diagnostic radiographs showed increased numbers of incipient carious lesions, so the patient's final treatment plan included stainless steel crowns on teeth #A, B, C, H, I, J, K, L, S, T to ensure full dental clearance for her bone marrow transplant (Figures 5-6). The patient recovered well and is now waiting for her bone marrow transplant.



Figure 5





DISCUSSION/CONCLUSION

Fanconi anemia is a medical condition that can greatly affect the dental treatment and modalities of pediatric patients. It is the most common cause of inherited bone marrow failure and cells cannot repair DNA damage, leading to genetic instability⁴. These patients are shown to have more recurrent infections, increased predisposition to malignancies, potential delayed and poor healing, and increased chance of hemorrhage¹. Malignancy predispositions mainly include acute myeloid leukemia (AML) and epithelial cancers of the head and neck³. There are many dental considerations to keep in mind when planning treatment for children with Fanconi amenia. Per this patient's hematologist, it is important to limit platelet transfusions for patients needing a bone marrow transplant, decreasing exposure to blood borne pathogens; this shows the importance of completing all treatment needed in one appointment or under general anesthesia if patient has lower than optimal platelet levels and is in need of a platelet transfusion prior to treatment. Due to increased risk of orofacial malignancies and oral manifestations of disease, routine follow ups and exams in pediatric patients is necessary. The most prevalent oral manifestations include gingivitis, periodontitis, recurrent aphthous ulcers, and eruption cysts¹. There is a potential increase in caries risk due to xerostomia from systemic effects of the disease¹. Fanconi anemia patients also present with an increased risk of leukoplakia and oral cavity squamous cell carcinoma, with these ailments affecting patients at younger ages with a likelihood of 500 times that of the general population^{2,4}.

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